Blastic plasmacytoid dendritic cell neoplasm: A rare subtype of myeloid leukemia

Hasmukh Jain *, Manju Sengar, Hari Menon, MV Chandrakanth

Hematolymphoid Unit, Medical Oncology, Tata Memorial Hospital, India

* Corresponding author. Address: Room No. 20, Ground Floor, Main Building, Tata Memorial Hospital, E. Borges Road, Parel, Mumbai 400012, Maharashtra, India. Mobile: +91 9167347482; fax: +91 22 2417 5505. · dr.hkjain@gmail.com · Accepted for publication 10 March 2014

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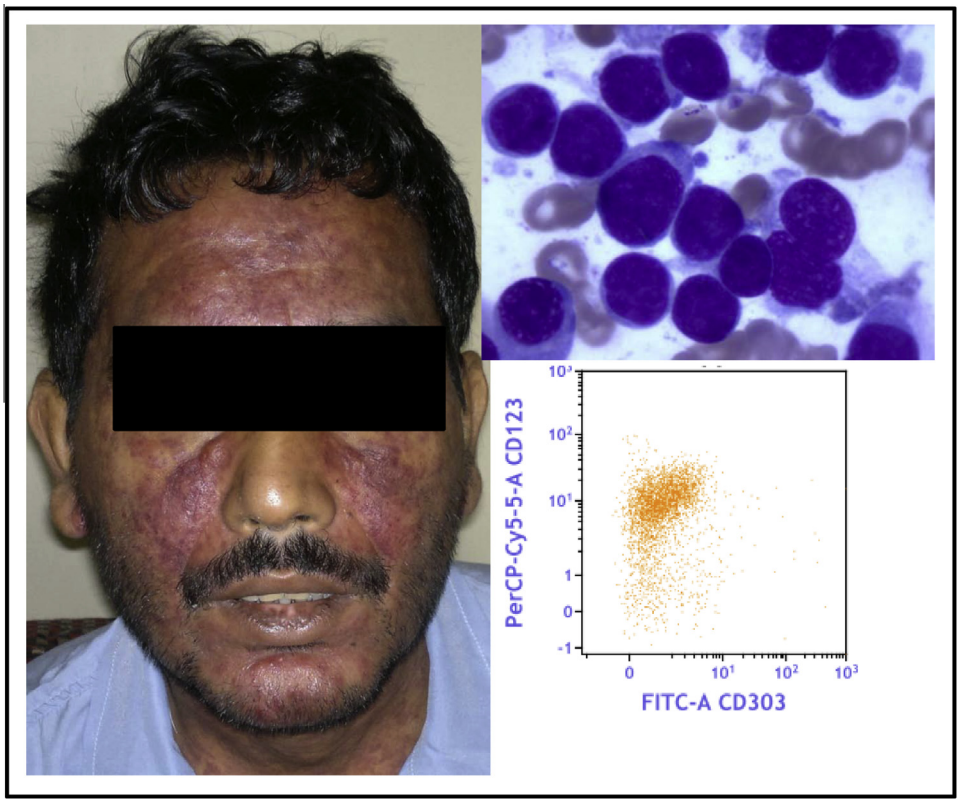
A 34-year-old male presented with a 2-month history of fever, backache, and generalized rash. Clinical examination revealed generalized lymphadenopathy along with maculopapular, erythematous, confluent rash involving the face, trunk, and extremities. Complete blood counts showed pancytopenia (hemoglobin 8.1 g/dL, platelet count 7000/mm^3, and white blood cell count 2000/mm^3). Bone marrow examination revealed intermediate to large sized blasts. The blast population expressed CD4, CD56, HLA-DR, dim to negative CD38 and CD45 and variable CD123. Blasts did not express CD 303, markers of myeloid, B-cell or T-cell lineage. The morphological and immunophenotypic findings were consistent with a diagnosis of blastic plasmacytoid dendritic cell neoplasm (BPDCN). BPDCN is a rare malignancy, and belongs to the category of acute myeloid leukemia and related precursor neoplasms. It commonly affects elderly males and involves skin, bone marrow and lymph nodes. The



blasts have abundant cytoplasm with a low nuclear-to-cytoplasmic ratio with microvacuoles beneath the nuclear membrane. They co-express CD4 and CD56 without common lymphoid or myeloid lineage markers. They may express CD 123, TCL1, CD 303, CD2AP, and the IL-3 receptor α-chain. Curative treatment options are limited and include acute lymphoblastic or myeloid leukemia-like induction chemotherapy followed by allogeneic stem cell transplant.

CONFLICT OF INTEREST

None declared.